Respiratory Medicine CME 4 (2011) 151–153

Contents lists available at ScienceDirect

Respiratory Medicine CME

journal homepage: www.elsevier.com/locate/rmedc

Case Report

A 42-year-old man with progressive dyspnea and a rapidly growing mass☆

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ARTICLE INFO

Article history:
Received 11 May 2011
Accepted 25 May 2011

Keywords:
Malignant fibrous histiocytoma
Mesothelioma
Pleural effusion
Undifferentiated pleomorphic sarcoma

ABSTRACT

Malignant fibrous histiocytoma (MFH), also known as undifferentiated high grade pleomorphic sarcoma, accounts for 20–24% of all soft tissue sarcomas in the US. Peak incidence is in the fifth and sixth decades of life, with a 2:1 male to female distribution. MFH is usually seen in the soft tissues of the extremity and involvement of the lung is rare. We report a case of pleural MFH in a patient with previous pleuro-pulmonary tuberculosis for which he had a lobectomy.

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1. Clinical summary

A 42-year-old white man presented with sub-acute onset of weight loss, malaise and pleuritic right sided chest pain. Three years ago he was diagnosed with pleuro-pulmonary tuberculosis for which he underwent a right upper lobectomy in addition to receiving anti-tuberculous therapy. He was an active smoker with a 40 pack-year history of cigarette smoking and drinks beer daily. He denied any illicit drug use or significant occupational exposures. Physical examination was relevant for diminished breath sounds with dullness to percussion posteriorly on the right lower lung zones. Laboratory testing was unremarkable. Chest imaging at presentation (Figs. 1 and 2) demonstrated a loculated right pleural effusion and right sided pleural thickening with nodularity. Thoracentesis revealed the pleural fluid to be an exudate with lymphocyte predominance. Smears, cultures and cytology were negative. Increasing pleural effusions and worsening respiratory status prompted a tube thoracotomy. His clinical condition deteriorated more and he developed respiratory failure requiring mechanical ventilator support. Repeat CT scan (Fig. 3) of the chest revealed rapid expansion of the tumor mass and persistent pleural effusion. A closed pleural biopsy was performed. Microscopic examination of the pleural biopsy (Fig. 4) revealed sheets of pleomorphic epithelioid cells with populations of intermingled spindle shaped cells. A high mitotic rate and marked necrosis were noted. Immunohistochemical staining profile demonstrated no definable line of differentiation. Electron microscopy did not demonstrate any microvilli. Given these findings, a diagnosis of malignant fibrous histiocytoma was made. Our patient subsequently developed ventilator associated pneumonia and septic shock. His tube thoracotomy continued to drain more than 1 L each day. He was clinically unstable for any aggressive surgical intervention. He had an unfavorable course, developing multi-organ failure and passed away 3 weeks following admission. Autopsy revealed a tan, 35 × 27 × 10 cm, fleshy, focally hemorrhagic mass encasing the right hemithorax (Fig. 5), from the apex of the chest cavity to the diaphragm inferiorly and to the anterior and posterior mediastinum medially. The tumor displaced the heart to the left by 5 cm. Arising from the pleura grossly, this tumor passed through the oblique fissure of the right lung. The previous upper lobectomy site, as evidenced by a surgical staple line, was also surrounded by the mass. Aortic adventitia, esophageal adventitia, pericardium and the crux of the diaphragm were all involved by the tumor. The cut surface of the tumor demonstrated soft, variegated, and pale-tan to hemorrhagic tumor parenchyma.

2. Discussion

MFH was first described by O'Brien and Stout[1] in 1964. MFH most commonly occurs in the soft tissue of the extremities, especially lower extremities.[2] Similar to other soft tissue sarcomas, malignant fibrous histiocytoma mainly metastasizes via the hematogenous route to the lung, liver, and bone.[2,3] While the lung is a common site for metastatic involvement of MFH, primary

Abbreviations: CD68, Cluster of Differentiation 68; CT, Computed Tomography; MFH, Malignant fibrous histiocytoma; MRI, Magnetic Resonance Imaging.

☆ Facility: The work described below was performed at the University of Louisville.

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1755-0017/$36.00 Published by Elsevier Ltd.
doi:10.1016/j.rmedc.2011.05.002

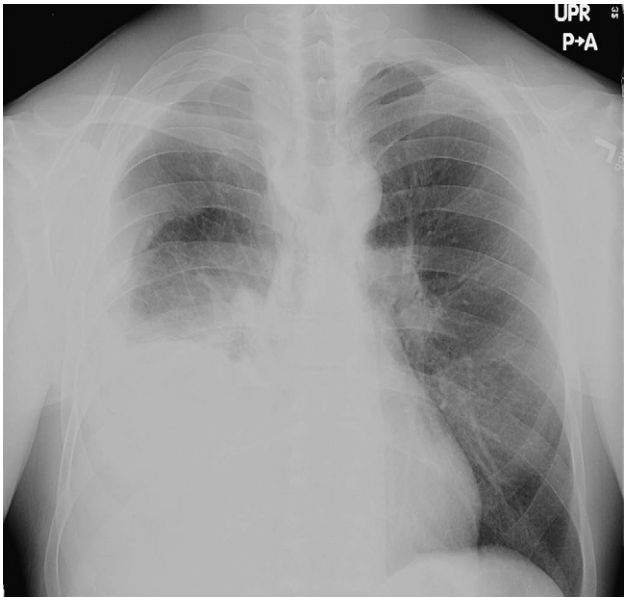

Fig. 1. Chest radiograph. Chest radiograph on presentation reveals opacification of right hemithorax with pleural thickening.

Fig. 2. CT scan of the chest (Initial). CT chest (with intravenous contrast) showing a large right pleural effusion with nodularity and thickening of the pleura.

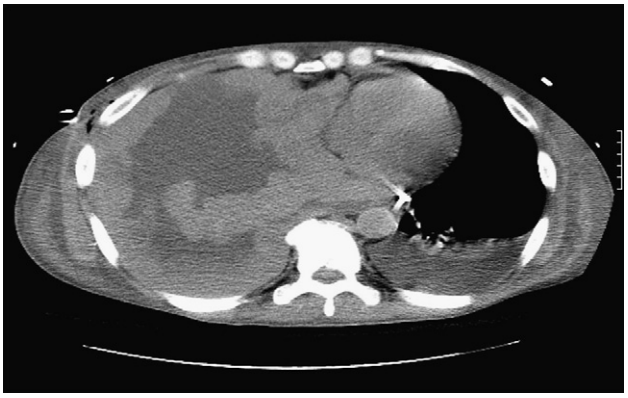

Fig. 3. CT scan of the chest (3 weeks subsequent to presentation). CT chest (without IV contrast) showing increasing thickening and nodularity of the pleura, along with the large pleural effusion.

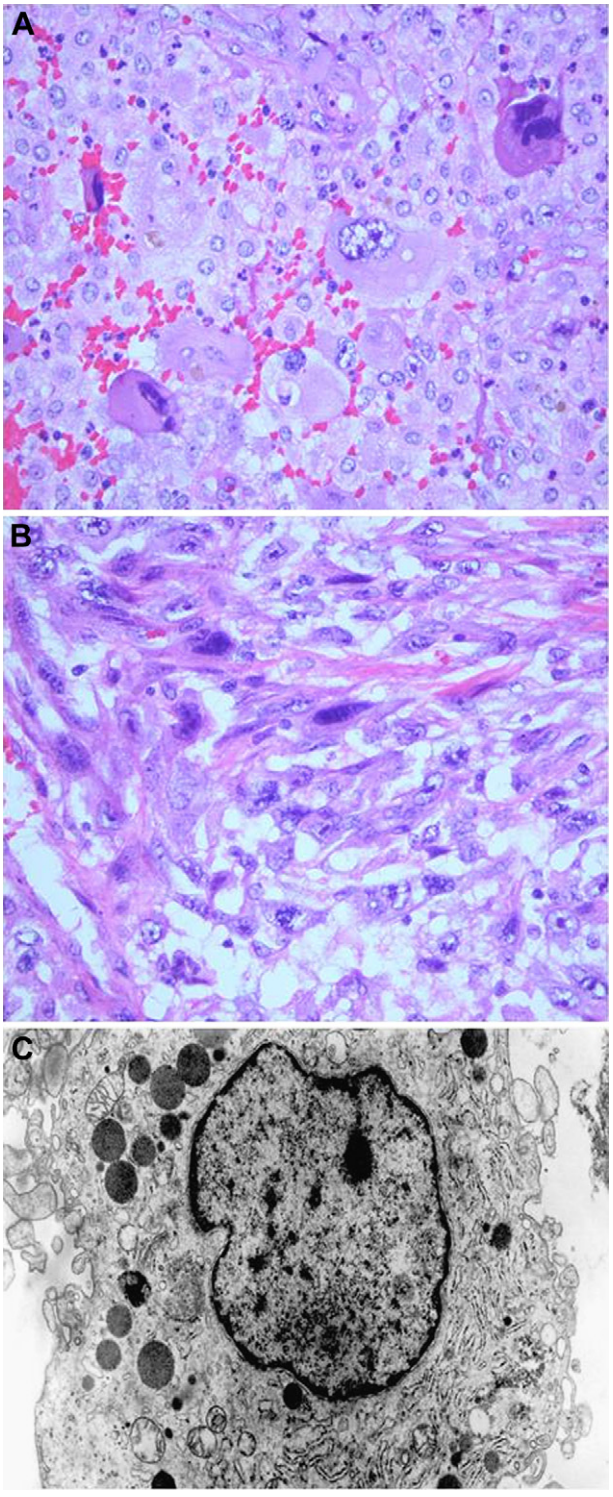

Fig. 4. Pleural biopsy. Hematoxylin and Eosin stained (A, B) pleural biopsy sections at 40x magnification showing pleomorphic spindles and epithelioid populations. Electron micrograph (C) showing cytoplasmic primary and secondary lysosomes and lack of villi on tumor cell surfaces (microvilli are specific for mesothelial cells).

involvement is rare, with approximately 60 cases reported in the English literature. Primary MFH of the pleura is rarer, with only 5 cases[4–8] reported in the English literature to date (Table 1). Risk factors include previous radiation exposure, shrapnel injury, pulmonary/pleural tuberculosis and implanted metallic devices. Common clinical presentation is a slowly enlarging painless mass,

Fig. 5. Gross pathology (Autopsy). Autopsy revealed a tan, 35 × 27 × 10 cm fleshy, focally hemorrhagic mass encasing the right hemithorax, from the apex of the chest cavity to the diaphragm inferiorly and to the anterior and posterior mediastinum medially.

Table 1
Reported cases of Malignant Fibrous Histiocytoma (English literature).

Age (yrs)	Sex	Tumor size	Survival[a]	Presenting symptoms	Pleural effusion	Comments
61	F	24 cms	2 yrs	Dyspnea	+	Hepatic metastasis noted 18 months after surgery[4]
62	F	[b]	2 yrs	Cough	+	Had cardiac tamponade due to tumor invasion[5]
65	M	12 cms	Alive 4 yrs follow up	Fever, fatigue	–	From initial evaluation, the mass was stable for 7 years, with increase in size and removal 8 years later[6]
51	F	8 cms	Alive 14 days post-surgery	Dyspnea, fever	+	Had a stable mass for 23 years, with rapid increase in size after that[7]
86	M	10 cms	13 yrs	Incidental	+	Slow growing mass noted when patient had surgical evaluation for peptic ulcer disease[8]

[a] Since initial tumor presentation.
[b] Not mentioned, circumferential pleural thickening was present with multiple nodules on the pleura.

with occasional reports of rapid expansion of tumor. CT scans typically reveal a large, lobulated, soft tissue mass of predominantly muscle density, with nodular and peripheral enhancement of solid portions. Central areas of low attenuation may be present, corresponding to myxoid regions, old hemorrhage, or necrosis. Fat attenuation is not observed in the tumors, which can distinguish tumors from some well-differentiated liposarcomas. MRI is useful when surgery is planned, helping to identify chest wall or pericardial involvement. The main treatment is wide margin surgical excision (extrapleural pneumonectomy) with or without adjuvant radiation or chemotherapy.

Diagnosis of MFH depends heavily on histology and exclusion of other more specific sarcomas by immunohistochemical stains and additional ancillary studies. MFH shows no definitive lines of differentiation. Microscopic examination of MFH demonstrates sheets of epithelioid appearing pleomorphic cells with eosinophilic or foamy cytoplasm and eccentrically located large nuclei. Typically a high mitotic rate and marked tumor necrosis are present. Immunohistochemical stains demonstrate a strong, diffuse intracytoplasmic staining for vimentin, alpha-1 antitrypsin, alpha-1 antichymotrypsin, CD68 and intense cytoplasmic staining for lysozyme. Electron microscopy reveals cells with histiocytic-like or fibroblast-like features showing pseudopodia with abundant primary and secondary lysosomes, ribosomes and some mitochondria. The nuclei of the tumor cells are large and irregular or indented. Both spindle and epithelioid shaped tumor cells are observed. The most important differential in this case is mesothelioma. Mesotheliomas stain positive for cytokeratin 5/6 and calretinin, as well as demonstrate microvilli on electron microscopy as compared to MFH.

3. Conclusion

MFH of the pleura is rare, with a varied presentation. It is usually a slow growing tumor and the histology does not correlate with its clinical behavior. The main prognostic factors are the degree of invasion of adjacent structures at the time of diagnosis. Complete excision is the treatment of choice, with long term follow up required evaluating recurrence.

Conflicts of interest

We have no conflicts of interest to disclose.

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